Interesting images

Gorham's disease: $^{99m}$Tc HMDP bone scan findings

Enfermedad de Gorham: hallazgos en la gammagrafía ósea con $^{99m}$Tc–HMDP

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A 45-year-old male was attended in the Emergency Department for pain in the left shoulder following effort. Physical examination did not show signs of functional limitation and radiology study was normal. Three months later the patient consulted for pain and edema in the left shoulder. Simple X-ray showed complete destruction of the humeral head with periarticular calcifications (Fig. 1a). A MR of the shoulder showed extensive tumoration centered in the glenohumeral articulation with well-defined borders, ruling out synovial sarcoma (Fig. 1b). On suspicion of a tumoral lesion a bone scan was requested for evaluation of extension.

The bone scan with 740 MBq of $^{99m}$Tc–HMDP showed abnormal uptake in the humeral neck, scapula, acromion and left glenoidal cavity. In the vascular and bone phase there was also an increase in activity in the soft tissues of the upper third of the upper left extremity (Fig. 2a and b). Low dose SPECT-CT showed that these lesions corresponded to calcifications localized in the adjacent muscular structures and important edema without visualizing the left humeral head (Fig. 2c). The diagnostic impression was neoplastic infiltration without ruling out pseudotumoral lesion (massive osteolysis of the humeral head) recommending

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**Fig. 1.** (a) Simple X-ray of the humerus: complete destruction of the humeral head with periarticular calcifications and (b) MR of the shoulder: extensive tumoration centered in the left glenohumeral articulation with well-defined borders.

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Fig. 2. Bone scan. (a) Vascular pool of the shoulders in anterior and posterior projection. Increase in the activity in the soft tissues of the upper third of the upper left extremity, (b) bone phase: whole body scan (anterior and posterior). Abnormal increased uptake in the proximal third of the left humerus and articular regions of the scapular girdle, with a defect in uptake in the humeral head, and (c) low dose SPECT-CT of the shoulders. The left humeral head is not visible. Abnormal uptake in soft tissues related to calcifications.

Fig. 3. Anatomopathological study. (a) Presence of vascular proliferation in relation to bone tissue (H&E, 4×) and (b) presence of osteoclasts in relation to bone tissue (H&E, 20×).

anatomopathological study. The bone biopsy confirmed osseous tissue with the presence of adjacent fibroconnective tissue with hypervascularization and hemangiomatous vascular proliferation without signs of malignancy (Fig. 3a and b) compatible with Gorham’s disease according to the clinical and radiological context.

Once diagnosed the patient started treatment with zoledronic acid and calcium plus vitamin D. The clinical evolution was favorable, with the patient remaining asymptomatic and with acceptable mobility. Inverse arthroplasty of the shoulder was proposed, but the patient is satisfied with his current mobility.

Gorham’s disease, also known as massive osteolysis or evanescent bone, is considered a rare disease of unknown etiology. It is characterized by local proliferation of small vascular or lymphatic channels which produce progressive bone destruction and spontaneous reabsorption of one or more bones around a focal point without respecting the articular limits.1,2 This disease may affect any part of the skeleton but is more frequent in the upper jaw, scapular girdle, ribs and pelvis1,2 and may present at any age, being more common in adolescents and young adults. Both sexes are equally affected and it is not associated with family history.1,2 Bone regeneration does not occur on the
cessation of the progression of osteolysis.\textsuperscript{3} The disease may be monostotic or polystotic, although multicentric involvement is exceptional.\textsuperscript{1,3} It is considered to be benign and osteolysis frequently stops after several years. The clinical presentation is variable, depending on the area affected.\textsuperscript{1} The diagnosis may be suspected or made following exclusion of other causes of osteolysis such as infection, tumor, inflammation or endocrine diseases and based on the combination of the clinical evolution, radiological findings and histopathological confirmation.\textsuperscript{1–3} Medical treatment includes radiotherapy, antosteoclastic medication (bisphosphonates) and alpha-2b interferon. Surgical treatment includes bone resection and reconstruction by bone graft or prosthesis.\textsuperscript{1,3}

References